

# Idiopathic aortitis with calcification of ascending aorta, and aortic and mitral valves<sup>1</sup>

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*A young woman with unexplained radiographic calcification of the ascending aorta was found at necropsy to have healed idiopathic aortitis. Calcification also involved the aortic valve which was stenosed and the mitral valve. Death was the result of infective endocarditis of these valves with aortic ring abscess, rupture of aortic root, and cardiac tamponade.*

Extensive radiographic calcification of the ascending aorta is infrequently encountered and has usually been attributed to syphilis (Higgins and Reinke, 1974). According to these authors the incidence of advanced forms of cardiovascular syphilis has decreased considerably in the past two decades, and other causes of aortic calcification now assume greater importance. In rare instances atherosclerosis (either primary or acquired on a bare area of a laceration of the aorta) may be associated with ascending aortic calcification (Levy *et al.*, 1963). Advanced atherosclerosis in elderly patients is the most frequent cause of such calcification at present (Higgins and Reinke, 1974). Two recent reports (McLoughlin *et al.*, 1974; Singleton and Merten, 1973) describe the unusual combination of extensive calcification of the ascending aorta and aortic valve in young women. The aetiology of these changes was unknown. A similar patient whose histology gives some insight into the basis for the calcification is reported.

## Case report

### Clinical features

The patient, a 24-year-old woman, was first seen at the age of 14 years with a history of mild spastic diplegia from childhood, and a cardiac murmur detected at the age of 8 years. She had no symptoms referable to her cardiovascular system.

Examination revealed normal peripheral pulses and a blood pressure of 145/100 mmHg (19.3/13.3 kPa) in

both arms. A grade 4/6 ejection systolic murmur was heard at the base with a soft early diastolic murmur. Electrocardiogram was normal, and extensive calcification of the ascending aorta, and aortic and mitral valves was seen on radiography. Cardiac catheterization revealed a 48 mmHg (6.4 kPa) peak systolic gradient across the aortic valve which had a calculated valve area of 0.7 cm<sup>2</sup>. The mitral valve functioned normally. Her Wassermann reaction and lupus erythematosus cell test were negative. Serum calcium, inorganic phosphate, and cholesterol were normal but total globulins were raised. The patient also suffered from psoriasis.

She remained asymptomatic over the following 10 years, with no alteration in physical signs, except that her blood pressure returned to normal. Left bundle-branch block with left axis deviation developed at the age of 15 years. She died at 24 years of age with the clinical features of infective endocarditis complicated by pericarditis.

### Pathological findings

Postmortem radiography of the heart and great vessels revealed calcification of the ascending aorta together with the aortic and mitral valves (Fig. 1). The arteries arising from the aortic arch were not calcified. The aortic valve had 3 cusps which were moderately thickened and rendered immobile by extensive calcification, resulting in aortic stenosis. The commissures were not fused. The aorta showed post-stenotic dilatation. The mitral ring was heavily calcified and the cusps showed mild thickening with scanty calcification.

Infected vegetations were present on the aortic and mitral valves (Fig. 2). Cardiac tamponade had followed rupture of the aortic root through an aortic ring abscess. The latter had also interrupted the bundle of His. A coagulase positive *Staphylococcus aureus* was cultured from the infection.

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FIG. 1 Postmortem radiograph of the heart and thoracic aorta showing calcification of the ascending aorta and left-sided heart valves.

The pulmonary and tricuspid valves appeared normal and there were no congenital cardiac anomalies. The coronary arteries arose from narrow, calcified aortic ostia. These vessels and their epicardial branches were devoid of atheroma. The pulmonary arteries appeared normal.

#### Microscopical findings

The major pathology was seen in the aortic media. The ascending aorta showed severe widespread medial destruction and calcification. In most areas the media had been totally destroyed and replaced by fibrous tissue, which had become calcified. Calcification was less severe in the arch where remnants of the original media in the form of fragmented elastic fibres separated by calcified collagen were present. Calcification of zones of medial damage about 20 elastic fibres thick was seen (Fig. 3). These lesions appeared as local areas of coagulative necrosis, with shrinkage of medial components. Scanty foci of intimal fibrosis were present. Calcification was medial in distribution and atherosclerosis was not important in this regard. No inflammatory cells were present. The adventitia showed fibrous thickening.

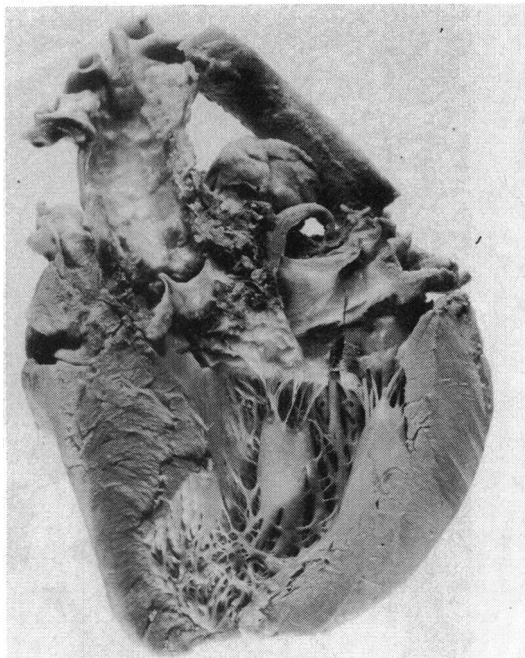


FIG. 2 Postmortem specimen showing calcified ascending aorta and left-sided cardiac valves. Infected vegetations are present on the aortic and mitral valves.

#### Comment

The microscopical findings in the aorta were compatible with the lesions of healed idiopathic aortitis (Takayasu's disease). Calcification of the aorta may occur in idiopathic aortitis (Hachiya, 1970), in the ascending aorta, the arch, and descending aorta. The unusual feature in our case was that the calcification involved the left-sided heart valves as well as the aorta. While aortic regurgitation (often associated with hypertension) has been recorded in idiopathic aortitis (Case records of the Massachusetts General Hospital, 1961; Judge *et al.*, 1962; Schrire and Asherson, 1964), we are unaware of any reported case with aortic stenosis or calcification of mitral valve. The cause of the valvular calcification in our patient is obscure.

There are striking radiographic similarities between our patient and the two cases each reported by Singleton and Merten (1973) and McLoughlin *et al.* (1974). Both of Singleton and

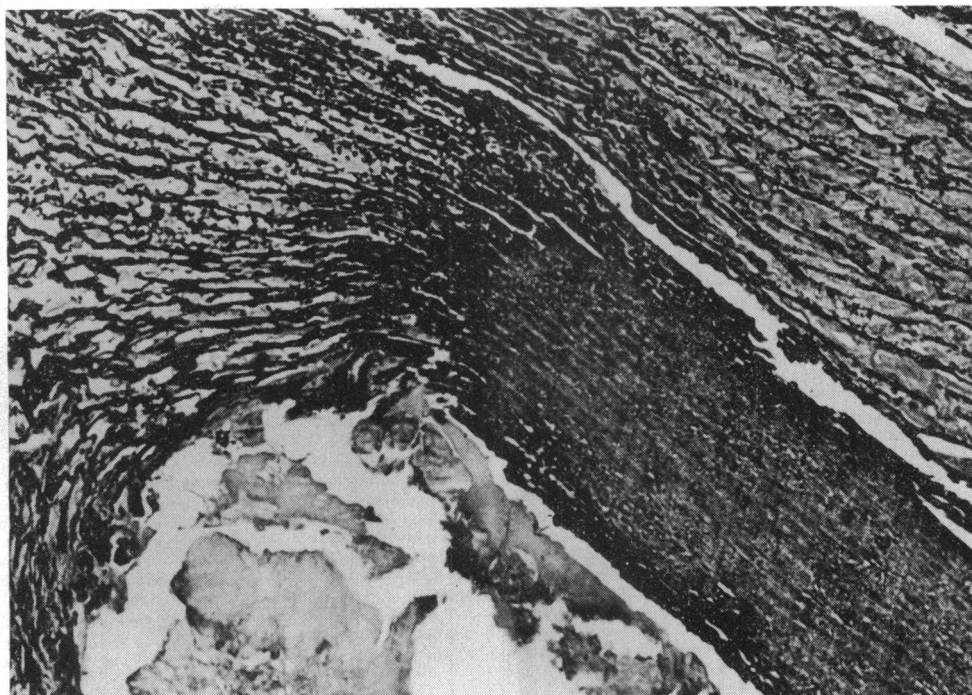


FIG. 3 Area of coagulative necrosis of aortic media external to paler staining area of medial calcification and fragmentation. (elastic van Gieson  $\times 115$ .)

Merten's patients had calcification of the ascending aorta and aortic valve (one had scanty mitral valve calcification). The 2 patients of McLoughlin *et al.* had calcification of the aortic valve and ascending aorta. The aetiology of these changes was unknown and aortitis was not considered. These 2 patients were still alive. Both patients described by Singleton and Merten had unexplained fevers at an early age. One of them showed hyalinized degeneration of the aortic media with focal calcification and localized hyperplasia of the intima over the calcified areas. The pathology was not illustrated, but the description sounds compatible, in our view, with healed idiopathic aortitis. Our patient illustrates that in idiopathic aortitis calcification may not only involve the ascending aorta but also the aortic and mitral valves.

### References

- Case records of the Massachusetts General Hospital (1961). Weekly clinicopathological exercises. *New England Journal of Medicine*, **264**, 664.
- Hachiya, J. (1970). Current concepts of Takayasu's arteritis. *Seminars in Roentgenology*, **5**, 245.
- Higgins, C. B., and Reinke, R. T. (1974). Nonsyphilitic etiology of linear calcification of the ascending aorta. *Radiology*, **113**, 609.
- Judge, R. D., Currier, R. D., Gracie, W. A., and Figley, M. M. (1962). Takayasu's arteritis and the aortic arch syndrome. *American Journal of Medicine*, **32**, 379.
- Levy, M. J., Siegal, D. L., Wang, Y., and Edwards, J. E. (1963). Rupture of aortic valve secondary to aneurysm of ascending aorta. *Circulation*, **27**, 422.
- McLoughlin, M. J., Pasternac, A., Morch, J., and Wigle, E. D. (1974). Idiopathic calcification of the ascending aorta and aortic valve in two young women. *British Heart Journal*, **36**, 96.
- Schrire, V., and Asherson, R. A. (1964). Arteritis of the aorta and its major branches. *Quarterly Journal of Medicine*, **33**, 439.
- Singleton, E. B., and Merten, D. F. (1973). An unusual syndrome of widened medullary cavities of the metacarpals and phalanges, aortic calcification and abnormal dentition. *Pediatric Radiology*, **1**, 2.
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